Medline

Cerebral amyloid in human prion disease.

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Neuropathol Appl Neurobiol 1993 Jun 19:253-60

BROWSE : Neuropathol Appl Neurobiol • Volume 19 • Issue 3
VIEW : **MEDLINE**, full MEDLINE, related records

Abstract

The clinical and neuropathological features of 21 patients with prion disease were reviewed with special reference to the morphology and immunoreactivity of cerebral amyloid. Six cases had a mutation at codon 102 of the prion protein (PrP) gene and in these the characteristic pathology was the formation of multicentric amyloid plaques which were stained with PrP antibody, whereas spongiform changes were absent in one and minimal in two. In one case, with a 216 base-pair insertion in the PrP gene, there was no spongiform encephalopathy (SE) but cerebellar amyloid was a prominent feature of the pathology. One case with a PrP gene mutation at codon 200 had severe SE but no amyloid. Two iatrogenic and 11 sporadic cases had SE and some form of amyloid was identified in all but three. Amyloid angiopathy and senile neuritic plaques, which stained with antibody to beta-protein, were present in familial as well as in sporadic cases, including some who were rather young to be regarded as having Alzheimer's disease. Cerebellar amyloid and degeneration of granule and Purkinje cells were particularly common findings in sporadic as well as in genetically determined cases. This study serves to emphasize the association between prion disease and amyloid deposition in the brain. PrP is a component of some amyloid plaques in a high proportion of cases with inherited prion disease but may also be found in cases of sporadic SE without known mutations or base-pair insertions in the PrP gene.

MeSH

Adult; Aged; Amyloid; Brain; Creutzfeldt-Jakob Syndrome; Female; Gerstmann-Straussler-Scheinker Disease; Histocytochemistry; Human; Kuru; Male; Middle Age; Neurites; Prion Diseases; Prions

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